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Congenital esophagostenosis due to tracheobronchial remnant in infant: 3 cases report☆

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ABSTRACT

Esophageal congenital stenosis due to tracheobronchial remnant is a rare deformation. Its scarcity incites us to report three observations of abnormal tracheal cartilage in the wall of the lower esophagus in infants (ages: 6; 12; 24 months). Authors present records of 3 children who underwent a radical procedure of surgery for esophageal congenital stenosis in Albert Royer's Pediatric Hospital from 2005 to 2009. Anatomopathological exam was performed to identify the cause of the stenosis and develop treatment recommendations. The study included two girls and one boy who presented vomiting, undernourishment and bad general state of health. The upper gastrointestinal radiological investigations established stenosis of distal esophagus with dilatation above. Endoscopy performed in one case showed not passable regular stenosis, located at 25 cm from dental arches. There was no abnormality of mucosal appearance above and dilation wasn't attempted for lack of pediatric dilator. Diagnosis of megaesophagus was formulated and all the infants underwent esophageal distal resection with terminal end-to-end anastomosis, HIS corner repairing and anterior partial stomach fundoplication to prevent reflux. The postoperative follow up was good and the upper gastrointestinal radiological investigations were normal after one month. The anatomopathological exam revealed tracheobronchial remnant in all cases. Authors discuss about standpoint to adopt in front of esophageal congenital stenosis due to tracheobronchial remnant. We believe that radical surgery is essential for the removal of stenosis.

Esophageal congenital stenosis due to tracheobronchial remnant is defined as the abnormal presence of a ring of tracheal or bronchial cartilage in the wall of the esophagus. It is an uncommon cause of congenital esophagostenosis among infants, which is usually revealed by vomiting when introducing solid food in early childhood [1]. Differential diagnosis is to be done with achalasia of the cardiac, fibro-muscular stricture and secondary stenosis especially gastro-esophagus reflux. The presence of cartilaginous ring has been proved to fail dilation and surgery appears as the unique treatment likely to remove completely and permanently the obstruction [2]. We report on three cases of infants in a four year period.

1. Methods

We reviewed the medical records of 3 consecutive infants who were admitted to our hospital with the diagnosis of megaesophagus. Gastrointestinal and esophagus Barium radiological investigation was performed in all cases. According to standard practice endoscopy examination is routinely required, but it was performed in one case as we had no easily available gastro-intestinal service support. The decision for surgical resection of the stenosis was made per-operatively and histological exam done on operative specimen. Satisfaction was based on recovery, vomit relief and weight gaining.

2. Results

2.1. Case 1

A 5-month 25-day-old female infant presents since she was 4-month-old, episodic early persistent vomiting. Then dehydration, severe undernutrition and moderate dyspnea were diagnosed following her hospitalization in pediatric unit with a weight of 3900 g in June, 28th 2004. On the 10th of July, face to persistent vomiting despite a well driven hydroelectrolytic resuscitation, an upper gastrointestinal radiological investigation was realized and showed stenosis of the distal quarter of the esophagus. On the 9th of September we performed gastrostomy of food supply with the aim of improving the nutritional state of the infant. The radical surgery decided 6 months later, on the 12th of January 2005,

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Fig. 1. Barium study.

consisted of a resection of the stenosed esophagus with an end-to-end anastomosis followed by an anterior partial gastric fundoplication. Postoperative consequences were simple and the histology results revealed a tracheobronchial remnant.

2.2. Case 2

A 9-month-old male infant without pathological history is admitted in pediatrics on September 17th, 2008 for early vomiting that runs since the age of 6 months in a context of moderate alteration of general state with an entrance weight of 8 kg. A correction of hydro-electrolytic disorders is made then an upper gastrointestinal radiological investigation is practiced on September 19th, 2008 showing a tight shrinkage of the terminal esophagus with a "socking" dilatation of the upstream segment (Fig. 1). Previously an endoscopy made on September 18th objectified a not passable, fibrous regular stenosis by the pediatric fiberscope located at 25 cm from dental arches; there was no abnormality of mucosal appearance above the stricture and dilation wasn't attempted for lack of pediatric dilator. The intervention is led by an upper umbilical median incision on December 24th, 2008 allowing resection of the terminal esophagus and end-to-end anastomosis with an anterior partial stomach fundoplication (Figs. 2–4). Postoperative consequences were simple and the hospital discharge was authorized after 8 days. The histological examination of the operative specimen found tracheobronchial remnant islet. After 24 months of follow-up the child is healthy and weights 12 kg.

2.3. Case 3

A 24-month-old presents, since the age of food diversification, early persistent vomiting in spite of usual antiemetic agents. She had been hospitalized on October 30th, 2008 in pediatrics in a picture of protein-calorie malnutrition with a weight of 8.400 kg.

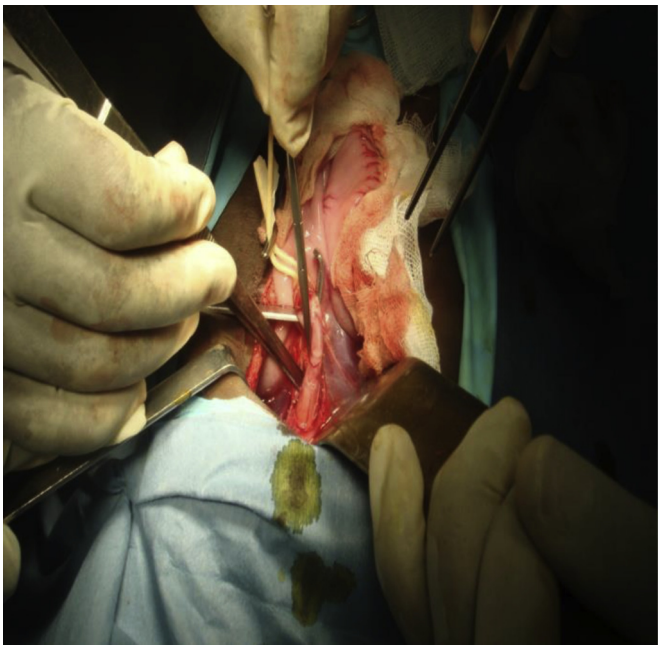

Fig. 2. Operative findings.

The upper gastrointestinal radiological investigation was made on November 17th, showing a regular shrinkage of the terminal esophagus surmounted by a "socking" dilatation of the upstream segment. The radical surgery was made on March 17th, 2009 with simple postoperative consequences and a weight recovery of 11.800 kg after a recession of 19 months. The histological results of the operative specimen revealed tracheobronchial remnant (Fig. 5).

3. Discussion

The prevalence of esophagus congenital stenosis revolves around 1/25,000–1/50,000 births according to Japanese studies [3]. This very rare affection is associated nearly in one third of the cases

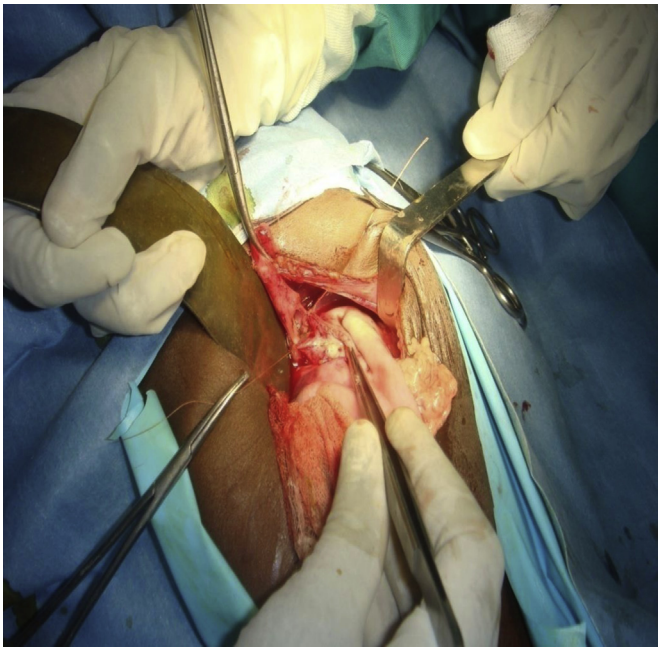

Fig. 3. End-to-end anastomosis after resection.

Fig. 4. Anterior partial stomach fundoplication.

with the other malformations (esophagus atresia, esophagus fistula) [4]. Tracheobronchial remnant constitutes a ratio of one sixth of esophagus congenital stenosis listed in the literature [5,6].

These sténoses are essentially found on the third of the lower part of the esophagus and preferentially the terminal esophagus [7]. Our study confirms that report and shows no note of peptic injury.

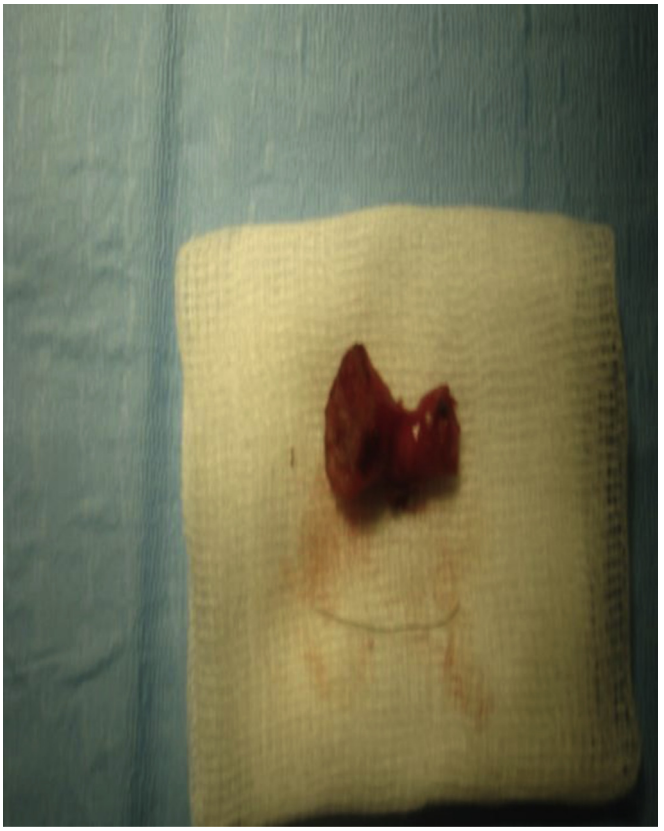

Fig. 5. Operative specimen.

The pathogenesis admitted by most of the authors seems to result of a defect of esotracheal separation. The phenomenon of vacuolization of the funiculus esophagus, between the fifth and the eighth embryonic week, is going to form the luminal tube. An intrauterine anoxia or the stress during this period can entail a disorder of the funiculus tubulisation leading to the stenosis. The sequestration of cells with tracheobronchial potential at the third lower part of the esophagus during the normal process of growth would explain the heterotopias [8].

The symptomatology is dominated by functional symptoms of persistent vomiting contrasting with a poor physical examination among the two oldest patients whereas for the 5-months-old infant the clinical picture came along with a severe undernutrition and respiratory distress syndrome. According to Vergos, vomiting, regurgitation and respiratory complications are the usual modes of revelation for the newborn while for the big child it is dysphagia the master symptom [9].

The diagnosis is based on the gastrointestinal radiological investigation showing a regular shrinkage well centered of the gastro-esophagus junction. Stenosis is surmounted in each case by a "socking" distension of upstream segment, feigning to be mistaken as an idiopathic megaloesophagus. In the endoscopic examination this stenosis appears generally in the form of a narrow path with a normal mucous membrane. However, the radiological and endoscopic aspects are not always characteristic and cast sometimes a doubt on the origin of the stenosis. The endoscopic ultrasound is another resource to be used in the attempt to identify the stricture of cartilage during surgery and allows removal or biodegradable stents [10,11].

The means of treatment are on the one hand instrumental by dilatations using balloon catheter and on the other hand surgical involving resection of the affected segment with an end-to-end esophageal anastomosis. Some authors perform circular myomectomy and stricturoplasty. Although there are reports of endoscopic balloon dilations as treatment for congenital esophagostenosis due to tracheobronchial remnant, it can result in serious complications such as perforation of the esophagus whereas radical surgery has low morbidity and mortality [12,13]. The evolution of the surgery about our patients was favorable in all cases with a recession of respectively 5 years, 2 years and 20 months. In the majority of the cases the medium and the long term evolution reported in the literature is very favorable after the surgery and the risk of recurrence is nil [14,15].

4. Conclusion

Congenital esophagostenosis due to tracheobrancheal remnant is a very rare deformation. Its diagnosis is often difficult but it is necessary to know how to think of it in front of vomiting or regurgitations arising in the introduction of solid food among children who have never been subject to vomiting during the exclusive breast-feeding. Recent publications on operative esophageal endoscopic ultrasound show that it plays a major role to distinguish fibro-muscular stenosis from stenosis due to cartilage. The surgical excision of the esophagus portion site of tracheobronchial remnants, by allowing complete recovery, remains the best treatment.

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